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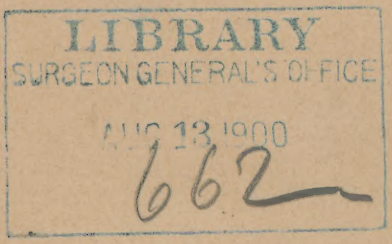
REPRINTED FROM THE JOURNAL OF CUTANEOUS AND GENITO-URINARY DISEASES, FOR JUNE, 1898.



[Reprinted from JOURNAL OF CUTANEOUS AND GENITO-URINARY DISEASES, June, 1898.]

CONGENITAL BULLOUS DERMATITIS WITH EPIDERMIC CYSTS.

By JOHN T. BOWEN, M.D.,
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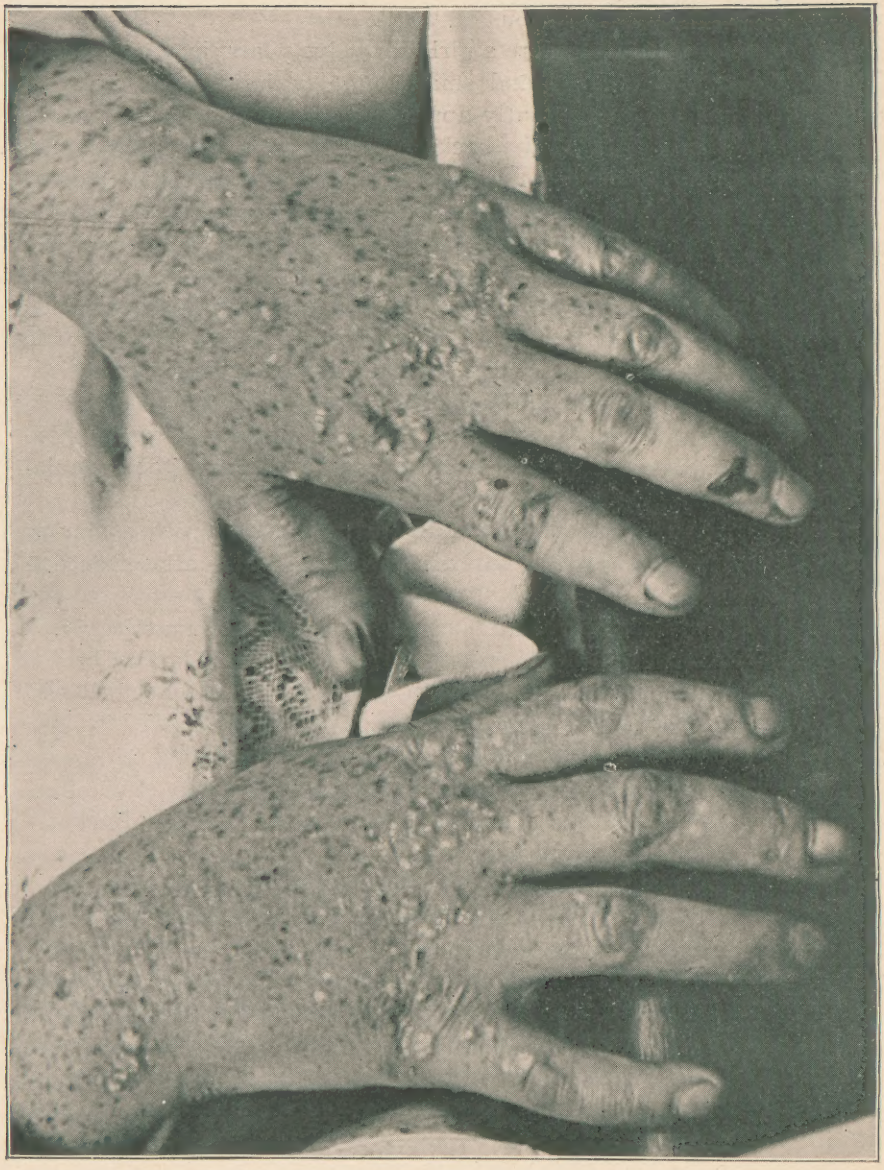
THE following case came under my observation during the summer of 1896:

The patient was a girl of twelve, born in America, whose parents were natives of New Brunswick. There was no history of any cutaneous affection in her family, and several brothers and sisters were living and healthy. According to the account of the mother, an extremely intelligent woman, the first signs of cutaneous disturbance appeared at the age of three weeks, when a "blister" containing fluid was observed on the dorsum of the foot, and since then there have been constant recurrences of bullæ, limited for the most part to the extensor surfaces of the legs, especially the knees and feet, to the elbows, and to the backs of the hands and wrists. These bullæ appear more frequently in summer than in winter, and are often produced by a slight blow or knock.

The patient was a thin and rather anemic child, with hair of a pronounced reddish hue. There were numerous large and small lentigines upon the face and hands. At the time of observation, which included several visits at intervals of a fortnight, the bullous eruption was most prominent upon the knees and the dorsal aspect of the feet. These parts were covered with areas of deeply reddened, slightly pigmented skin, not very sharply bounded from the normal integument, and somewhat infiltrated. These hyperemic

and thickened areas had obviously been produced by the long-continued, recurrent eruption of bullæ in these places. At present these areas are found to be covered with bullæ, some of the size of a pea, others with a diameter of from half an inch to an inch, which rise abruptly from the hyperemic and thickened skin. The fluid in some of the bullæ is clear, in many others it is of a dark hemorrhagic color, as seen through the translucent wall. There was one very large hemorrhagic bulla on the outer aspect of the lower leg, otherwise the front of the knees and the dorsum of the feet were the only parts below the waist affected. The trunk was absolutely free from lesions. The elbows were affected in the same manner as the knees and feet, *i.e.*, there were areas of hyperemic, pigmented skin upon which were situated bullæ. In this situation the bullæ were mostly of small size, and usually filled with clear fluid, and were not so numerous as upon the knees and feet. The backs of the hands also presented a few bullæ, not very numerous at the time of observation, but according to the mother's testimony these regions had been repeatedly the seat of very marked outbreaks. The most prominent appearances on the hands were firm, whitish nodules, resembling milia. They varied in size from a pin's head to a small pea. They were most numerous over the knuckles where they showed a marked tendency to arrange themselves in groups, and in some places, as shown in the photograph, a distinct circular arrangement was seen, the edge of the circle being made up of these small white nodules, while the center was either free or occupied by one or two lesions only. They were present, also, to a less extent upon the backs of the wrists and upon the backs of the fingers, especially over or near the articulations. There were also a number of lesions on the thick skin of the palms, seated upon large reddened areas, similar to those described upon the knees, feet, and elbows. When these milia-like bodies are squeezed a soft, cheesy substance is given out. The nails of the fingers are striated longitudinally and brittle. The nails of the toes are brittle, crumbly, and broken. In the scalp there are a few doubtful crusted lesions, and on the side of the neck a reddened, thickened area of the diameter of an inch, covered with the milia-like bodies, but without any appearance of bullæ at the present time. The mucous membranes are intact, but the mother says that at a number of times there have been lesions in the mouth. In the areas occupied by the bullæ there are slight superficial cicatrices, evident only on close inspection, and therefore not well brought out in the photograph. On the face there have been from time to time a few bullæ, but no milia-like bodies are to be seen here, and there is no

FIG. 1.



scarring. The trunk, upper legs, and arms, with the exception of the elbows, have never been affected. There is considerable pruritus accompanying the eruption, but the child is of an exceedingly ner-

vous, fretful temperament. The mother, without being questioned, volunteered the information that a knock or blow would often be speedily followed by a bullous outbreak.

To sum up, we have here a girl of twelve years, in whom, since three weeks of age, large and small bullæ, with often hemorrhagic contents, have been constantly appearing symmetrically on certain regions of the body, *viz.*, the backs of the elbows, the backs of the wrists, hands, and fingers, the anterior aspect of the knees, and the dorsum of the feet. Occasional lesions have also appeared on the face, mucous membrane of the mouth, palms of the hands, and the anterior aspect of the lower legs. These bullæ are often produced by a slight knock or blow. In the regions where these bullæ have repeatedly occurred in great numbers the skin is intensely hyperemic and somewhat thickened, and there is slight but still perceptible scarring. In these areas, the seat of repeated bullous outbreaks, bodies closely resembling milia are seen. These have a tendency to group and form circles, and are most in evidence over the articulations of the hands and fingers.

This case at once brought vividly to mind those with a similar train of symptoms reported by Hallopeau, of which a brief outline may be given.

At the first meeting of the French Society of Dermatology and Syphilis, in 1890, the first case presented was by Hallopeau, under the title, "Une dermatose bulleuse congénitale avec cicatrices indélébiles, kystes épidermiques et manifestations buccales." The patient, a boy of seventeen, had suffered from the age of six weeks from constantly recurring attacks of bullæ, which were often filled with blood, and appeared symmetrically on certain parts of the body only, *viz.*, the extremities, elbows, and neck; they were also present on the mucous membrane of the mouth. They were often surrounded by an erythematous areola and were succeeded by cicatrices and small epidermic cysts in the form of miliary nodules. At one period the eruption had covered the body generally. There was much deformity of the nails, some of which had been lost. A microscopic examination of the miliary nodules by Darier showed that they were in fact epithelial cysts, seated in the upper part of the corium and probably due to the obliteration of the sweat-gland outlets and the follicles by a proliferation of connective tissue consecutive to the bullous dermatitis. Unfortunately, Darier was unable to trace a complete connection between the epithelial cysts and the underlying glands.

Hallopeau at the same time spoke of a case with similar appear-

ances that had been shown by Vidal the previous year. In this case, a girl of thirteen, the eruption had made its appearance during the first month of life. Besnier observed that bullæ filled with blood were sometimes seen to make their appearance under the action of a very slight bruise, or of any kind of an irritant, and that permanent scars were the common result. He had never seen a case where these phenomena were congenital, however. These cases had been described under the name of pemphigus.

In 1896 Hallopeau[1] again referred to the first case reported by him. The patient had remained under observation and had continued to have the same bullous eruption with hemorrhagic contents. Hallopeau had satisfied himself that the bullæ were often produced under the influence of light knocks. He then described a second case that he had just seen. The patient was a man of fifty-five who had been affected with the eruption since his birth. It consisted of constantly recurring outbreaks of bullæ with hemorrhagic contents, on the elbows, knees, backs of the feet, hands, fingers, and toes. The nails were prominently affected, and the bullæ were succeeded by scars and by the same milia-like nodules as in the other case. The outbreaks were almost constant, the slightest knock or bruise being sufficient to cause them to appear. An interesting feature of this case was that the patient's great grandmother, grandmother, and mother had had the same eruption limited to the same localities, and, furthermore, that the patient's son, at present in military service, was also affected in the same way. Within the last two months the eruption had become more generalized, appearing over the lower legs, about the axillæ and umbilicus, and on the back and thighs. There was also much pruritus, which had not been a feature previously.

In commenting on this case Hallopeau lays great stress on the remarkable symmetry that the eruption had assumed in the latest outbreak, where it had followed the distribution of the nerves. This fact, together with the mode of development of the bullæ under the influence of a slight knock or blow, he considers conclusive proof of the trophoneurotic nature of the eruption. Hallopeau recognizes the resemblance of these cases to the affection described by Goldscheider, Köbner, and Blumer, under the name of "hereditary tendency to the formation of traumatic bullæ or epidermolysis bullosa," and by Valentine as "hereditary bullous dermatitis." He considers, however, that the cases of which we have been speaking are to be distinguished from those described by Goldscheider, Köbner, and

[1] Hallopeau, *Annales de Derm. et de Syph.*, p. 453, 1896.

Valentine, by the inflammatory character of the eruption, by its seats of predilection on the dorsal aspect of the articulations, by the fact that the soles of the feet were not affected, by the cutaneous atrophy and scarring, by the epidermic cysts which followed the bullous outbreaks, by the development of bullæ without appreciable traumatism, and by the occurrence of acute outbreaks following the distribution of the nerves.

At a meeting of the Société Française de Dermatologie et de Syphiligraphie of June 3, 1897, Augagneur describes another case with a somewhat similar train of symptoms. In his case the person affected was a man of sixty-four; the affection was congenital and had the same seats of predilection—elbows, knees, and dorsal aspect of the articulations. The affected areas were transformed into a cicatricial reddened surface, resembling that which follows superficial burns. There were no epidermic cysts. In this case, also, an attack was observed when nearly the whole body was affected, accompanied by great pruritus. The bullæ were usually produced by a slight trauma. Augagneur believes that there are not sufficient grounds to warrant a separation of this and like cases from the epidermolysis bullosa hereditaria of Köbner. The constant feature is the production of bullæ by insignificant traumatism, and the preservation of the general health. Inconstant features are heredity, the spontaneous appearance of the bullæ, the complete disappearance of the disease in certain cases, and the formation of epidermic cysts.

Under the title "Epidermolysis Bullosa," Wallace Beatty[1] has collected a large number of published cases of congenital bullous eruption produced by traumatism. He reports three cases of his own, of a man and his two children. The man, who was forty-one years of age, had all his life been subject to the occurrence of bullæ, with sometimes clear and sometimes hemorrhagic contents which appeared especially on the elbows, knees, hands, and feet, as well as the scalp. On his knuckles he used to have groups of small yellow-white points, like those seen in his children, but he does not have them now. The skin over the areas that have been repeatedly affected was much reddened, loose, shiny, and wrinkled; one small milium was present in a patch of thickened and reddened skin below the left knee. The nails were either absent or imperfectly formed.

The elder child, a girl of three years, had been subject to the occurrence of bullæ since two months of age, when they first appeared on the hands. Since they have recurred constantly under the prov-

[1] *British Journal of Dermatology*, August, 1897.

ocation of a slight blow or bruise, on the elbows, knees, wrists, and hands, especially about the knuckles, and the feet. There are groups of "milia" over some of the phalangeal joints of the fingers and a few in the palms. There were a few small milia on the back of the neck, a place where there had never been bullæ. The nails were slightly affected. The younger child, a boy of eighteen months, was born with a bulla on the right thumb. There are reddened areas and bullæ on the same localities as in the sister's case, *viz.*, knees, elbows, hands, and feet, especially the extensor surface of the phalanges, and in the latter situation there were groups of milia. The milia-like bodies were examined microscopically and showed cysts seated just below the epidermis, of which the wall was made up of a narrow layer of epithelial cells, while the cavity was partly empty and partly made up of horny masses of cells that did not stain. A direct continuity with the epidermis could not be traced, and therefore their mode of origin was not determined. There could be no doubt that they were of epithelial origin, but their presence on the palms of the hands where there are no hair-follicles or sebaceous glands, showed that they were not necessarily connected with these structures.

Beatty also describes several cases under this same category that had been published under other headings, notably two cases reported by Wickham Legg in 1883 as congenital pemphigus persistent from birth. All of these cases, including those of Hallopeau, Legg, and his own, Beatty considers to belong in the same class with epidermolysis bullosa, as described and illustrated by Goldscheider, Köbner, Blumer, Elliot, and others. Beatty gives a full *résumé* of all the published cases of epidermolysis bullosa.

In the *British Journal of Dermatology* for September, 1897, Colcott Fox illustrates the difficulties to be met with in our attempts to evolve a little order out of the present chaos of bullous eruptions. His title is "Pemphigus in a Woman of Nine-Years' Duration, at First Indistinguishable from Ordinary Pemphigus; afterward with All the Clinical Characteristics of Pemphigus Congenitalis (Epidermolysis); Epidermic Cysts; Essential Shrinking of the Conjunctiva." The patient, a woman of fifty-four, had been first attacked nine years previously, just before she had become pregnant with her youngest boy. The eruption was extensive and severe, so that she was twice admitted to the hospital. At this time there was no grouping or localization of the lesions. After the pregnancy the affection gradually assumed a chronic course with especial features. It was found that the bullæ arose from very slight injuries, such as from pressure

of the corsets, at the place where the teeth come into contact with the tongue and lips, and on the parts exposed to knocks and bruises, as the elbows, knees, hands, and feet. There were also bullæ on the mucous membrane, and there was a condition of so-called "essential shrinking of the conjunctiva." At one time there were numerous epidermic cysts on the regions that had been repeatedly affected, but at present these have disappeared. The skin on these seats of predilection had gradually become atrophic and cicatricial, so that it somewhat resembled an old scleroderma. The nails of the hands and feet had been permanently lost from the repeated occurrence of bullæ beneath them.

Fox asks if this case is to be grouped with epidermolysis. The only objections are that it did not begin until middle life, that it was like an ordinary pemphigus at the beginning, and that there was no family tendency. The latter objection can hardly be called such, as the rule of heredity has not yet been shown to be absolute in this affection. He considers that further experience is necessary before this question can be decided. Fox also thinks it an error for Hallopeau to separate his cases of congenital bullous dermatitis from epidermolysis. He does not think the points of differentiation referred to by Hallopeau sufficient to warrant an independent place for this group of symptoms. Epidermic cysts may occur in other types of pemphigus and may be consecutive to a simple burn.

In our attempt at classifying the bullous eruptions included heretofore under the vague and meaningless term "pemphigus," we should, as Dr. Fox asserts, tread cautiously. The difficulties in our path are very great, but it is to be admitted that in the last ten years a certain amount of progress has been attained. The cases of epidermolysis bullosa, or hereditary tendency to the formation of traumatic bullæ, as described by Goldscheider, Köbner, Blumer, Elliot, and others, are certainly entitled to an individual place, as they apparently have no relation with most other conditions of so-called pemphigus. We have here a congenital and often hereditary vulnerability of the skin to such a degree that a very slight injury, chiefly in the form of friction or pressure, causes bullæ to appear on the parts of the body so affected. The name epidermolysis given to the affection by Köbner is not appropriate, since according to Elliot's investigations we have to do with a true inflammation of the skin; a view that is also sustained by Unna.

When we come to consider the case reported at the beginning of this article we see that it corresponds exactly with those described by Hallopeau under the title "Dermatose bulleuse congénitale avec

cicatrices indélébiles, kystes épidermiques et manifestations buccales." Beatty's own cases and several of those enumerated by him in his article on "Epidermolysis Bullosa" have the same combination of symptoms. I must agree, however, with Augagneur and Fox that neither the scarring nor the epidermic cysts renders to these cases the right to a separation from the class of congenital bullous dermatoses, in which epidermolysis bullosa is included. Bodies undoubtedly identical in nature with these cysts have been found in other forms of bullous eruption. Hebra refers to a case related by Von Bärensprung where a very extensive crop of "milia" appeared over the greater part of the body of a girl who was suffering from chronic pemphigus. He relates, also, a case of his own where numerous small white nodules, thickly clustered in groups and circles, and in every way similar to milia, appeared on the dorsal and flexor side of the wrist and on the dorsal aspect of the fingers and toes in a case of recovery from pemphigus—localities where there had been previously pemphigus bullæ. It must be admitted, however, that the description is not sufficiently detailed to exclude the possibility that one or both of these cases belonged in the class of congenital bullous dermatitis that we have been describing, although it would not appear so.

Handford[1] reports the case of a girl of thirteen, who suffered for a year from a bullous eruption which was preceded by a polymorphous erythema, the bullæ appearing at the site of the erythematous lesions. The eruption appeared in crops with a high temperature, occasionally 104° F. When admitted to the hospital there was a bullous eruption in all stages of healing, chiefly on the buttocks and flexor side of the thighs and knees, on the ankles and toes, and also the extensor surfaces of shoulders, elbows, and wrists, and the dorsum and palms of the hands. There were bullæ on the mucous membrane of the mouth. The bullæ left smooth *pigmented cicatrices* on healing. About five months after the beginning of the affection the skin of the hands, wrists, feet, and ankles was red and cicatricial and covered with groups of opaque white spots, which had almost disappeared when the patient left the hospital. These bodies were found to consist of degenerated epithelial cells. Handford remarks that the partial limitation to the extensor surfaces of the upper extremities, the back of the shoulders, and inside of feet and ankles, suggests mechanical injury and irritation as an important cause. There is a plate added that illustrates the milia-like bodies upon the foot. It is to be noted that when the patient passed out of observation at

[1] "Tr. Clinical Socicty," London, 1888.

the end of a year, although she was much improved, isolated bullæ were still appearing.

This case has perhaps certain points of similarity with that of Fox, that has just been referred to. For how long the bullæ contin ued to form on the parts exposed to pressure and injury in Handford's case is not known; but we have here repeated outbreaks of bullæ, accompanied by high temperature. Later the process assumed a more chronic phase, and the occurrence of bullæ was chiefly limited to the parts of the body most exposed to injury, where a cicatricial condition of the skin, and the appearance of milia-like nodules followed the bullæ. It is certainly difficult to assign a place to either of these cases.

The cases of Handford and Fox, however, show conclusively that scarring and the appearance of epidermic cysts may follow bullous eruptions that are not congenital. It is to be noted, however, that in both of these instances the eruption had as its seats of predilection the parts most exposed to injury, and that both of the writers speak of this as a probable exciting cause. Again, cases occur similar in other respects to those of Hallopeau and the one described by me, where scarring and the formation of epidermic cysts are not present. In September, 1896, I saw a child of eight, whose eruption, according to the mother's story, had begun at the age of two years. It consisted of successive outbreaks of bullæ limited to the extensor surfaces of the elbows, wrists, knuckles, knees, and feet. There were also some lesions on the buttocks and thighs. The affected areas were reddened, thickened, and pigmented, but there was no scarring and no appearance of epidermic cysts. The nails were not affected.

I am in perfect sympathy with Colcott Fox, who urges that we should, with our present imperfect knowledge of etiological conditions, proceed cautiously in our attempts at subdividing the group pemphigus. We may fairly, however, set apart most if not all of the cases that have been referred to here, and it will conduce to simplicity and further advancement in our knowledge, if the term pemphigus, as far as they are concerned, is dropped. Congenital bullous dermatitis seems preferable to the term epidermolysis bullosa as being less prejudicial to any further knowledge of the etiology. It should be kept in mind that under this heading belong the cases described by Goldscheider, Köbner, Blumer and others, of hereditary tendency to the formation of traumatic bullæ, where bullæ arise repeatedly under the influence of often very slight traumatism; and also (possibly as a separate group) the cases to which especial refer-

ence has been made in this article, where the seats of predilection are the extensor surfaces of the joints, and where the additional features of atrophy and the formation of epidermic cysts, usually, but not always, are present.